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Case Report

Granular cell tumor of anal border

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ABSTRACT

The objective of this report is to describe a case of granular cell tumor of the anal border and to review the most relevant topics of the literature on the subject. Ours is a female patient, 57 years old, with an asymptomatic nodule in the anal border for 2 years. Surgical excision was performed, with a histopathological diagnosis of granular cell tumor. The first description of this tumor was carried out in 1926 by Abrikossoff. The techniques of immunohistochemistry and electron microscopy allowed us to determine its origin in Schwann cells. These are rare tumors, most often diagnosed between the 4th and 6th decade of life and, in general, are benign formations – only 2% of them are malignant. These tumors can occur in any part of the body, although they are more common in the oral mucosa, dermis and subcutaneous tissue. The treatment solely by surgery has a curative effect, and its recurrence is unusual. The location in the anal/perianal area occurs even more rarely, and we found only 48 cases previously described in the literature.

Tumor de células granulares de borda anal

Palavras-chave:
Canal anal
Tumor de células granulares
Neoplasias
Relatos de casos

RESUMO

O objetivo deste relato é descrever um caso de tumor de células granulares de borda anal e revisar os tópicos mais relevantes da literatura acerca do tema. Trata-se de uma paciente do sexo feminino, 57 anos, com histórico de nódulo na borda anal assintomático há 2 anos. Foi realizada ressecção cirúrgica da lesão, com diagnóstico histopatológico de tumor de células granulares. A primeira descrição deste tumor foi em 1926 por Abrikossoff. As técnicas de imunohistoquímica e de microscopia eletrônica permitiram determinar a sua origem nas células de Schwann. São tumores raros, mais frequentes entre a 4ª e 6ª década de vida e, no geral, benignos, apenas 2% são malignos. Podem ocorrer em qualquer parte do corpo, embora sejam mais comuns na mucosa bucal, derme e tecido celular

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subcutâneo. O tratamento cirúrgico isolado é curativo e a recorrência incomum. A localização no ânus/canal anal/perianal é ainda mais rara, sendo encontrados apenas 48 casos previamente descritos na literatura.

Introduction

The first histopathological description of the granular cell tumor was performed in 1926 by Abrikossof. These are rare tumors, predominantly benign. The occurrence of malignant granular cell tumors barely reaches 2%, requiring observation of histological features of malignancy and evidence of metastases for its diagnosis.[1,2]

These tumors can show a wide distribution throughout the body, although the most affected areas are the buccal mucosa, dermis and subcutaneous tissue. Granular cell tumors are more frequent between the 4th and 6th decade of life.[3] In most case series, the tumor prevails among males, ranging from 64.5% to 68%.[2,4] Isolated surgical treatment is curative and its recurrence is unusual and as a general rule occurs in the same local, being associated with an incomplete resection of the primary lesion.[2–4]

Here we report the case of a granular cell tumor of the anal margin and discuss the most relevant aspects of literature and those presented by this case.

Case report

Female patient, 57 years old, African descent, teacher, married. A previously healthy patient without clinical and surgical comorbidities and no relevant family history; complained of an asymptomatic nodule located in the anal margin for about two years. She denied changes in her bowel habits and in characteristics of evacuations. On examination, a hardened perianal formation measuring about 2 cm, in the 5.00 o'clock position, was observed.

Surgical excision was indicated. Preoperative evaluation with colonoscopy, chest X-ray and laboratory tests found no abnormalities. Thus, the entire macroscopic lesion was resected at surgery.

In the macroscopic pathology, two surgical fragments of mucosa, with 3.2 cm × 1.3 cm × 1.6 cm and 1.3 cm × 0.7 cm × 0.4 cm, respectively, were evaluated together. The microscopy revealed an anal mucosa lined by stratified squamous epithelium without atypia, displaying typical irregular acanthosis features in the epithelium adjacent to the tumor. In the corium, we found cell proliferation with a large and granular cytoplasm, with regular nuclei, forming small groups amid intense fibrosis (Figs. 1 and 2). There was no necrosis or significant mitotic activity. The process was compromising the adjacent adipose tissue and skeletal muscle, in addition to lateral and deep surgical margins. The immunohistochemistry was positive in the neoplastic cells to S100, CD 68 (Figs. 3 and 4), neural specific enolase, inhibin and calretinin; and negative for pan-cytokeratin (AE1 and AE3).

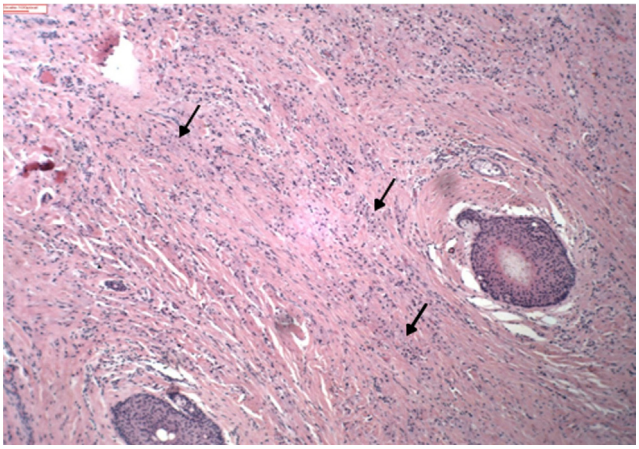

Fig. 1 – Tumor cells, low magnification, hematoxylin-eosin staining (40×).

Imaging studies (abdominal computed tomography and magnetic resonance imaging of the pelvis) carried out after surgery showed only postoperative changes in the anal area.

Discussion

Virtually, granular cell tumors can occur in any anatomical location, and although more cases are located in the skin and subcutaneous tissue, the tongue is singly the most common site. In 5–8.5% of the cases, the tumors are multiple. Rarely these formations are diagnosed before the surgical excision or when obtaining a biopsy of suspicious lesions.[2,4,5]

The presence of this tumor in the gastrointestinal tract varies from 5% to 19%.[2,4] Among the tumors of the perianal

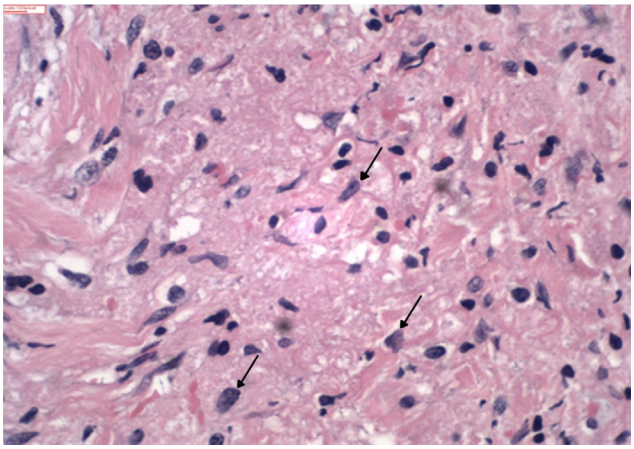

Fig. 2 – Tumor cells, large magnification, hematoxylin-eosin staining (400×).

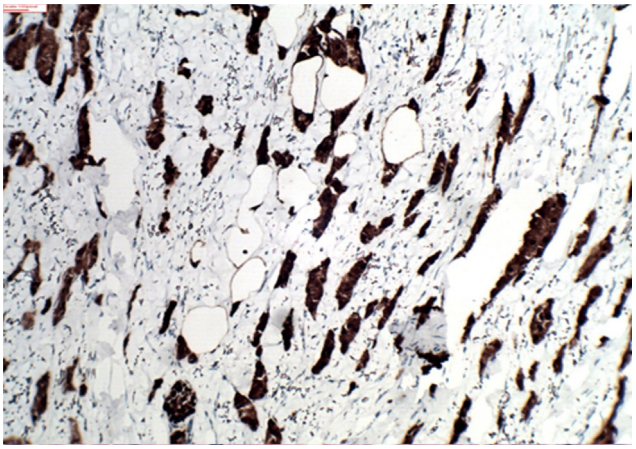

Fig. 3 – Immunohistochemistry: groups of positive tumor cells for S100 protein (100×).

region, there may be rectal bleeding; but most of them present as painful nodules that may be confused with abscess. Many of these formations are diagnosed incidentally during an evaluation of other changes, such as hemorrhoids and perianal fissures.[6,7]

Among the described histopathological features, one can highlight the positivity in immunohistochemistry for S-100 protein and CD-68. The S-100 protein is expressed in various tumors (neural tumors, melanomas, sarcomas of clear cells, histiocytosis, and others), and the positivity, in the case here presented, supports the neural origin of the lesion, as it is currently accepted in the literature. Despite this origin, the tumor cells share many morphological and structural characteristics of macrophages; thus, these cells are positive for CD-68, a macrophage/histiocytic marker par excellence. Another important aspect is the frequent association of these tumors with some degree of pseudoepitheliomatous hyperplasia of the overlying epithelium, which can be a confounding factor with squamous cell carcinoma.[4,6]

Malignant granular cell tumors are rare (2% of cases).[1,2] Some histological characteristics favoring the diagnosis of malignancy are: the presence of necrosis, a vesicular nucleus with an obvious nucleolus (in benign tumors, the nucleus is pyknotic), a trend for the formation of spindle cells, and some mitotic activity. Some authors suggest the classification of tumors in those with uncertain malignant potential, if these formations have any of the above characteristics, besides favoring a more rigorous clinical follow-up and the survey for occult metastases.[1]

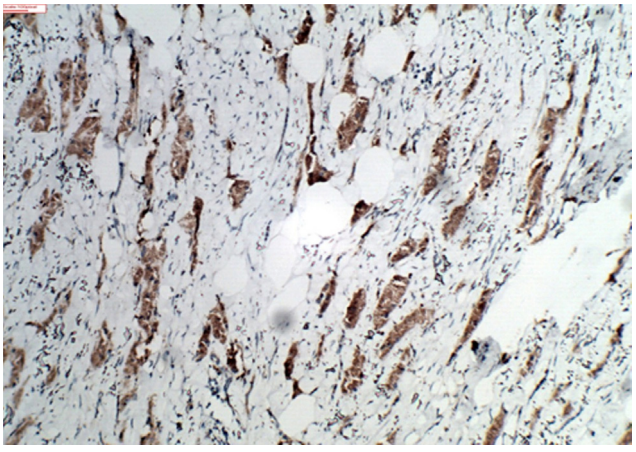

Fig. 4 – Groups of positive tumor cells in immunohistochemistry for CD-68 (100×).

Malignant tumors affect (more than benign tumors) deep soft tissues, esophagus, larynx, peritoneal cavity and peripheral nerves. The major metastatic sites are lymph nodes (70%) and lung (50%). Malignant variants have an aggressive behavior, with a rate of local recurrence of 70%, and result in death in 65% of cases after an average of 2.5 years after diagnosis. Among the differential diagnoses of malignant granular cell tumors, alveolar soft tissue sarcoma and rhabdomyosarcoma can be particularly cited and, less commonly, dermatofibrosarcoma, malignant fibrous histiocytoma and melanoma.[1]

Literature series

Lebranchu et al. published the largest series of granular cell tumors, with 263 cases, of which 15 were multiple and 12 were associated with malignancies of other histological features, but with no cases of malignant granular cell. There was only one case of recurrence after an incomplete resection. Men were affected in 68% of the time and the main sites were: skin (38%), esophagus (19%) and tongue (10%). Nine cases occurred in the digestive tract (21.03% of total) and only 1 in the rectum and 1 in the anus[2] (Table 1).

Lack et al. reviewed 118 tumors in 110 patients and found a higher prevalence of African descent (29%) patients, 64.5% in men, 44% located in the skin and subcutaneous tissue. The predominance of men was attributed to a possible selection bias of patients who came from military hospitals. Only six cases were located in the rectum/anus, which is the most common site in the gastrointestinal tract, followed by esophagus (2 cases) and stomach (1 case) (Table 1). In this series, recurrence was 8%; and in all cases, the surgical margin was compromised in the first resection. However, the absence of recurrence was numerically higher, even in cases with a positive surgical margin.[4]

The largest number of tumors in the perianal region was reported in a series only of the gastrointestinal tract: 16 cases in a total of 74. In total, only 48 cases of granular cell tumors in the perianal region have been described in the literature, highlighting the scarcity of the tumor in this site. Among these cases, 63.3% (19/30) occurred in male subjects, and the mean and median ages were 40.9 years and 39 years, respectively[6] (Table 1).

In conclusion, the present case shows clinical and pathological features similar to those described in the literature. This is a rare and benign tumor and the importance of this report reinforces the fact that its surgical treatment in the perianal location can result in morbidity and damage to the quality of life of patients, when the surgery results in fecal incontinence. For these reasons, despite the compromised surgical margins, the best option in the present case may be a regular follow-up, with reconsideration of a surgical approach only in the case of tumor recurrence.

Table 1 – Description of cases of granular cell tumors located in the rectum/anal canal in the literature.

Author and year of publication	N° of cases and location	Gender/age
Crane, 1945[8]	1, perianal	F/26
Simon, 1947[9]	1, perianal	F/50
Rothchild, 1953[10]	1, perianal	M/36
Rosenberg, 1960[11]	1, perianal	M/38
Winne,1961[12]	1, perianal	M/26
Vance, 1969[13]	6, perianal	(–)
Strong, 1970[5]	2, perianal	(–)
Rickert, 1978[14]	3, perianal	M/64
		M/39
		F/57
Lack, 1980[4]	3, rectal	(–)
	3, anal	
Johnston, 1981[6]	16, anal	M/25
		M/39
		M/23
		M/44
		M/31
		M/41
		F/50
		M/26
		M/24
		F/22
		F/32
		M/51
		M/25
		F/52
		F/44
		(–)
Pruglo, 1985[15]	1, rectal	M/33
Lisato, 1995[16]	1, rectal	(–)
Lebranchu, 1999[2]	1, rectal	(–)
	1, anal	
Nakachi, 2000[17]	1, rectal	F/47
Cohen, 2000[18]	1, anal	F/75
Al Bouzidi, 2003[19]	1, anal	M/56
Kim, 2003[20]	1, rectal	M/49
Santoni, 2006[21]	1, anal	F/28
Mourra, 2011[7]	1, anal	M/50

(–), information not available; M, male; F, female.

Conflicts of interest

The authors declare no conflicts of interest.

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